CHAPTER III.5. COST OF CARDIAC ABNORMALITIES

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CHAPTER III.5. COST OF CARDIAC ABNORMALITIES

III.5.A Background

This chapter contains a discussion of the methods used and the results of estimating the direct medical costs incurred by individuals with cardiac abnormalities and the results of the analysis.[1] It does not include information on elements such as indirect medical costs, pain and suffering, lost time of unpaid caregivers, etc. The reader is referred to Chapter I.1 for a discussion of the cost estimation methods and cost elements that are relevant to all benefits estimates. In addition, Chapter III.1 contains information regarding the special characteristics of developmental defects, and a list of chemicals that may cause developmental abnormalities.

The costs presented in this chapter were current in the year the chapter was written. They can be updated using inflation factors accessible by clicking on the sidebar at left.

Link to Chapters I.1 and III.1
Link to inflation factors

III.5.A.1 Description

A number of cardiac anomalies occur at birth or in early infancy, and are quite varied. These are structural defects in the development of the heart, arteries, and associated tissues. They arise when the function, movement and relationships among cardiac cells fail to progress normally. Five defects, all conotruncal heart anomalies, are discussed in this chapter. The specific anomalies include: truncus arteriosus, transposition of the great arteries, double-outlet right ventricle (DORV), single ventricle, and tetralogy of Fallot. They were selected from among the cardiac abnormalities for which cost data are available because they occur with greater frequency than other defects and are not usually fatal when treated. Each anomaly is described below in turn in Section A, followed by the cost data in Section B.[2]

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[1] "Costs" in this chapter refers to direct incremental per capita medical costs, unless otherwise noted.

[2] Certain forms of DORV are grouped together for the cost analysis because they resemble transposition and actually fall under the same International Classification of Diseases ICD-9 codes.

III.5.A.2 Causality

Information on causality is discussed for each specific cardiac anomaly described below. Many anomalies occur concurrently with other cardiac or non-cardiac anomalies. Although there appears to be a genetic component in some anomalies, none of the anomalies described below has a strong clear hereditary pattern. The causation of the anomalies is therefore for the most part unknown. These anomalies may be due to a variety of factors, including maternal health, heredity, and environmental factors.

A recently completed study of the offspring of pesticide application workers found that their incidence of circulatory system anomalies was significantly greater than that of the general population in the same area of the United States (Garryet al., 1996). Respiratory system, musculoskeletal, and urogenital anomalies also occurred at increased rates. The chemicals evaluated in the study that were associated with the birth defects were trifluralin, triazine herbicides (including atrazine, a very common well contaminant in agricultural areas), and chlorophenoxy herbicides (including MCPA and 2,4-D, a pesticide with very high usage).   There was also a significant increase in birth defects among infants conceived in the spring (i.e., during peak chlorophenoxy use), compared to infants conceived during other periods of the year (Garry et al., 1996). As indicated in the introductory developmental effects chapter (III.1), the timing of exposure is often a critical determinant of whether and what type of effects will occur.

Generally, the earlier during a pregnancy that damage occurs, the more serious the effect will be, because all cells developing from the damaged cells may also be damaged or eliminated, and basic structures are being formed during the first trimester (three months). Although Gerry et al. (1996) do not provide detailed information on the nature of the circulatory and other anomalies, information can be obtained from the author or from EPA (which funded the work).

Table III.1-1 in Chapter III.1 lists numerous chemicals associated with developmental abnormalities in human and/or animal studies. Many of these studies have identified structural and anatomical defects. Cardiac abnormalities fall into this category of defects.

Link to Table III.1-1 in Chapter III.1

III.5.A.3 Truncus Arteriosus

III.5.A.3.1 Description

Truncus arteriosus occurs when abnormalities in major arteries and valves lead to a mixing of oxygenated and de-oxygenated blood. This mixing results in an insufficiently oxygenated supply of blood to tissues. This disorder ultimately leads to congestive heart failure if uncorrected. In the absence of surgery, 75 percent of children die within the first year, and the survivors have severely limited abilities due to heart disease (Oldham et al., 1972). Truncus arteriosus occurs in approximately two percent of children with congenital heart defects (Oski, 1994).

III.5.A.3.2 Concurrent Effects

Children with this defect also frequently have underdevelopment of the aortic arch, displacement or stenosis at the origin of the coronary arteries, and absence of the main pulmonary artery supplying the lungs. This and other cardiac anomalies occur with greater frequency among Down syndrome children (statistics on occurrence were not available) (Waitzman et al., 1996).

III.5.A.3.3 Treatment and Services

Although previously considered an inoperable defect, substantial improvements in surgical and other treatments have been made in recent years. The surgical methods and medical follow-up continue to be refined (Oski, 1994). This defect is usually surgically treated at 6 to 12 weeks through constructive surgery and insertion of valves. Follow-up surgical repair or valve replacement is often required within the first five years after surgery (15 to 50 percent). All valves are replaced at twelve years due to growth. Follow-up surgery (i.e., dilations) is not uncommon (Waitzman et al., 1996).

III.5.A.3.4 Prognosis

Based on a small sample (106 infants) there is a six percent mortality rate prior to surgery and a ten percent mortality rate during the surgical period (Ebert et al., 1984; Bove et al., 1989). Complications may result from follow-up medical treatments (Waitzman et al., 1996).

III.5.A.4 Transposition of the Great Arteries

III.5.A.4.1 Description

Transposition of the great arteries occurs when the major arteries are transposed, leading to insufficient oxygen flow to tissues and lungs. This condition causes widespread cyanosis (Oski, 1994) and is fatal without surgical intervention (Waitzman et al., 1996).   Transposition of the great arteries is a common cardiac abnormality, occurring in approximately five percent of all patients with congenital heart disease (Oski, 1994).

III.5.A.4.2 Concurrent Effects

Various associated cardiac abnormalities involving multiple structures within the heart complicate the treatment of this disorder (for a detailed description see Oski, 1994). Ventricular septal defect also occurs in 20 percent of children with this anomaly (Kirklin, 1991).

III.5.A.4.3 Treatment and Services

Surgery to correct this defect is performed shortly after birth. Ten percent of affected individuals require additional surgery within one year (Waitzman et al., 1996). As noted under concurrent effects above, concurrent heart defects often occur, and the constellation of effects is considered when determining the appropriate medical treatment.

III.5.A.4.4 Prognosis

During the immediate post-surgical period, the mortality rate is approximately 14 to 22 percent, depending on the specific nature of the defects and procedures used. Between 30 and 80 percent of children with this defect who have received medical treatment develop cardiac arrhythmias within ten years, again depending on the defects and surgical approaches. Three to ten percent of these children require insertions of pacemakers (Waitzman et al., 1996).

Cardiac dysfunction occurs in approximately ten percent of children within ten years. Arrhythmias and heart failure lead to death in approximately 11 percent of children between 30 days and 10 years after surgery (Morris and Menashe, 1991). Depending on the type of procedures done to correct this disorder, various medical problems may arise, some requiring surgery at a later date. For a detailed description of the common long-term sequelae, see Oski (1994).

III.5.A.5 Double-outlet Right Ventricle

III.5.A.5.1 Description

Double-outlet right ventricle refers to a diverse group of heart defects that occur when both the aorta and pulmonary artery originate from the right ventricle, and a septal defect is present (Oski, 1994). This condition leads to insufficient blood flow to critical tissues, and may cause cyanosis, exhaustion with exercise, heart failure, and pulmonary vascular disease (Waitzman et al., 1996). This disorder occurs in approximately two percent of congenital heart defects. Although sometimes associated with trisomy-18 and material diabetes, most cases occur with no other congenital anomalies (Oski, 1994). The symptoms of this anomaly vary with the specific structural defects and may include cyanosis, congestive heart failure, and related effects.

III.5.A.5.2 Concurrent Effects

Complex lesions of the atrial or ventricular septum, valve defects, mislocation of the heart, and ductus arteriosus have also been observed with this defect (Waitzman et al., 1996).

III.5.A.5.3 Treatment and Services

Surgery is usually performed at 6 to 24 months. Arrhythmias in children are common, even with surgery, and may require follow-up care (Waitzman et al., 1996).

III.5.A.5.4 Prognosis

Various mortality rates have been reported in the literature. A 15 percent post-surgical mortality rate in infants was reported by Judson et al., (1983). Shen et al. (1990) reported that approximately 25 percent of children who have received appropriate medical attention die during childhood, probably due to arrhythmias. More recent reports indicate a much better prognosis with a 90 to 95 percent *post*-surgical survival rate and excellent functional status following surgery (Oski, 1994).

III.5.A.6 Single Ventricle

III.5.A.6.1 Description

Single ventricle occurs when one, rather than the usual two ventricles are present. One may dominate (they are usually balanced) and the other may be very small, or there may be only one present. In either case, there is one that is primarily functional (Oski, 1994). This anomaly causes insufficient blood flow to tissues and can lead to cyanosis, heart failure, and pulmonary vascular disease. There is a 70 percent mortality rate in childhood in the absence of surgery (Waitzman et al., 1996). This disorder is found in approximately one percent of children with congenital anomalies.

III.5.A.6.2 Concurrent Effects

Multiple concurrent cardiac defects occur with this anomaly. Twenty to 40 percent of children also have other non-cardiac problems, including scoliosis and lack of a spleen (Waitzman et al., 1996).

III.5.A.6.3 Treatment and Services

Correction of this defect commonly involves two surgical interventions, one shortly after birth, and one at 18 to 36 months. Additional surgery is required in 14 percent of children (Waitzman et al., 1996).

III.5.A.6.4 Prognosis

With surgery, survival varies considerably. The mortality rate is 29 to 43 percent within ten years. Among survivors, ten percent have limited activity and three percent were severely limited (Waitzman et al., 1996). Most patients exhibit exercise intolerance and cyanosis. Causes of death include dysrhythmia, congestive heart failure, brain abscess, pancreatitis,

cerebral infarction, cerebral embolus, and hemorrhage, and pulmonary embolus and valve occlusion (Oski, 1994). Due to the long-term medical problems associated with this anomaly, research into improvements in surgical and other medical treatments is being carried out. There is not a single currently accepted treatment method at this time, and various approaches are used. (Oski, 1994).

III.5.A.7 Tetrology of Fallot

III.5.A.7.1 Description
Tetrology of Fallot refers to a group of abnormalities of the heart that have the common characteristics of unrestrictive ventricular septal defects and an obstruction of the right ventricular outflow (Oski, 1994). These abnormalities also involve malpositioning of the aorta and thickening of the right ventricular wall (Pinsky and Arciniegas, 1990). The severity varies considerably from a heart murmur to life-threatening hypoxia. If it is not detected during infancy it may lead to a misshapen (boot-shaped) heart (Oski, 1994). This disorder occurs in approximately six percent of children with congenital heart defects (Oski, 1994).

III.5.A.7.2 Concurrent Effects
Multiple additional cardiac defects are associated with this anomaly Waitzman et al., 1996). Non-cardiac abnormalities are associated with this disorder in approximately 16 percent of cases, more so than with most other cardiac defects. The children are also more likely to have concurrent effects that are more serious than those found with other cardiac defects, including cleft lip and palate, hypospadias (reproductive organ abnormality in males), and skeletal malformations (Oski, 1994).

III.5.A.7.3 Treatment and Services
When Tetralogy of Fallot is detected shortly after birth, surgery to correct the defects usually occurs at three to twelve months of age. Depending on the specific nature of the defects, surgery may also be required shortly after birth. Five to fifteen percent of children require additional surgery within thirteen years (Waitzman et al., 1996). Treatment of arrhythmias may be required in some patients long after surgery, and bacterial endocarditis may also occur and require treatment later in childhood or adulthood (Oski, 1994).

III.5.A.7.4 Prognosis
The rate of post-surgical mortality is three to eight percent in infants (Touati et al., 1990; Walsh et al., 1988). At ten years the survival rate is 87 to 90 percent, and 85 percent survive into their twenties. The prognosis for normal function and activity is good in surviving children (Waitzman et al., 1996). As noted under the treatment section above, a number of problems may be anticipated in the long-term follow-up of children with this disorder (e.g., bacterial endocarditis, arrhythmias) and effects of

coronary artery disease at a more advance age may be more severe in people with this disorder (Oski, 1994).

III.5.B Costs of Treatment and Services

III.5.B.1 Methodology

Waitzman et al. (1996) provide an estimate of the direct medical and non-medical costs of treating cardiac abnormalities, specifically for the five types listed above. For the purpose of the cost analysis, transposition and double-outlet right ventricle (DORV) were grouped together. Waitzman et al. used the same methodology to estimate the costs incurred by individuals with each type of cardiac abnormality as for all the birth defects for which they estimated costs. The methodology and relevant considerations are detailed in Chapter III.3, including discussions of direct and indirect costs, prevalence versus incidence, incremental costs, and concurrent effects. The analytic method, the sources of data, and the limitations of the Waitzman method are also discussed in Chapter III.3. The methodology is outlined briefly here.

Link to Chapter III.3

To estimate the lifetime medical costs incurred by an individual with a birth defect, Waitzman et al. estimated the average lifetime medical costs for an individual with the birth defect. From this value, the authors subtracted the average lifetime medical costs for an individual without the birth defect. Because they estimated lifetime costs, they used an incidence-based approach. Ideally, they would have tracked the costs of the cohort members over time, until the death of the last cohort member. Because the members of the cohort were born in 1988, however, this tracking was not possible. Instead, estimates of the costs incurred at each age were based on estimates of per capita costs in the prevalent population of that age (see Chapter III.3, Section III.3.B.1.2).

Link to Chapter III.3, Section III.3.B.1.2

This method has two important implications. First, Waitzman et al. estimated the costs incurred by individuals with birth defects, including all medical costs incurred, rather than the cost of the birth defect per se. These cost estimates therefore include the costs of concurrent effects (unlike the costs reported for many of the diseases in this handbook). This method yields a more comprehensive assessment of total costs than would be obtained if only individual effects were evaluated. This method is of particular use in valuing the avoidance of birth defects because they very frequently occur in clusters within an individual. As Waitzman et al. note, however, the costs of associated anomalies are included as part of the

estimate of the costs incurred by an individual with a given birth defect. These cost estimates therefore cannot be aggregated across birth defects because of the possibility of double counting.

Second, the Waitzman et al. method estimates the *incremental* costs for individuals with birth defects — that is, the costs above and beyond the average costs that would be incurred by individuals without the birth defect.

Waitzman et al. (1996) estimated three categories of costs incurred by individuals with limb reductions: direct medical costs, direct nonmedical costs, and indirect costs.[3] Direct medical costs, specifically inpatient care, outpatient care, pharmaceuticals, laboratory tests, X-rays, appliances, and long-term care are included in the cost estimates shown in this and other chapters (Chapters III.3 through III.8) based on the work of Waitzman et al. Nonmedical direct costs, specifically developmental services, and special education are also included in this handbook.

The Waitzman estimates of the costs incurred by individuals with limb reductions are based on the costs of this birth defect in California across many ages, and its occurrence in a large cohort of children born in California in 1988. California's ongoing birth defects monitoring program provides an excellent source of data. The California data sets were linked with other national data sets so that Waitzman et al. could estimate the incremental costs associated with each type of cardiac abnormality.

The method of calculating the expected lifetime incremental costs for an individual with a birth defect — i.e., the average lifetime cost per case — is the same for all the birth defects considered by Waitzman et al. The expected per capita cost at age i, PCC_i, for an individual born with the birth defect is the probability of surviving to age i (among those individuals born with the birth defect), ps_i, times the per capita cost among individuals who do survive to age i ($PCPREV_i$, measured in the prevalent population):

$$PCC_i = (ps_i) \times (PCPREV_i) .$$

Waitzman et al. estimate per capita costs in the prevalent population of age i, $PCPREV_i$, in two different ways, depending on data availability (see Chapter III.3).

Link to Chapter III.3

The present discounted value of expected per capita lifetime costs of the birth defect, PCCOBD, is just the sum of these expected age-specific per

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[3] Indirect costs are not generally discussed in this handbook and so are not included in this chapter. The reader may wish to consult Waitzman et al. (1996) for information on these costs.

capita costs, appropriately discounted (as explained more fully in Chapter III.3):

$$PCCOBD = \sum_i PCC_i/(1+r)^i .$$

III.5.B.2 Results

Waitzman et al (1996) estimate the total lifetime medical costs of each of the cardiac defects according to the methodology outlined above. As outlined in Table III.3-1 in Chapter III.3, the prevalence on July 1, 1988 and the incidence in 1988 of cardiac abnormalities in the state of California was as follows:truncus arteriosus: 1,591 and 56, respectively; transposition/DORV: 7,469 and 263, respectively; tetralogy of Fallot: 5,336 and 187, respectively; and single ventricle: 1,932 and 68, respectively.   The occurrence data provide an indication of the relative rate of yearly occurrence of the different anomalies discussed in this handbook.

Table III.3-1 in Chapter III.3

Table III.5-1 shows the annual per capita medical costs incurred by individuals with each type of cardiac anomaly by age group with costs updated from 1988 to 1996 dollars based on the medical care cost component of the Consumer Price Index (1996:1988=1.6465).

Table III.5-1: Annual Per-Capita Medical Costs of Heart Defects Patients by Age Group (1996$)				
Condition	**Age 0-1**	**Age 2-4**	**Age 5-17**	**Age 18+**
Truncus arteriosus	$162,463	$104,225	$7,197	$5,139
Transposition/DORV	$56,296	$15,790	$3,071	$1,744
Tetralogy of Fallot	$70,729	$18,952	$5,939	$1,251
Single ventricle	$57,119	$15,727	$12,867	$10,071

The medical cost of the average population was then subtracted from these costs to obtain incremental costs. Waitzman et al. (1996) discounted these costs using three different discount rates: two percent, five percent, and ten percent. Although these discount rates do not match the standard EPA rates used in many other chapters in this handbook (zero percent, three percent, five percent, and seven percent), there is insufficient information provided in Waitzman et al. (1996) to allow a conversion to discounted costs using standard EPA discount rates. This problem exists in all chapters based on the Waitzman et al. data (i.e., Chapters III.3 through III.8).

The present discounted values of average per capita lifetime incremental costs, using discount rates of two percent, five percent, and seven percent, are listed in Table III.5-2 below. Direct medical costs and direct non-medical costs are listed separately. The sum of per-capita direct medical and nonmedical costs provides an estimate of the total per-capita costs incurred by individuals with each type of cardiac abnormality.

Children with some heart anomalies have a much lower probability of survival (see the "prognosis" descriptions in part III.5.A, under the specific descriptions above) than children with most cardiac or other anomalies discussed in this section of the handbook. This report focuses on medical costs, whereas other research often includes estimates of the value that a person would pay to save his or her life, termed "value of life" estimates. In the case of heart anomalies, survivorship is an issue for some of these children. The cost estimates presented here could therefore be dwarfed by the additional value of life associated with each estimate.

Table III.5-2: Per-Capita Net Medical Costs, Nonmedical Costs, and Total Costs of Heart Defects (1996$)			
Cost Element	2%	5%	10%
Truncus arteriosus			
Net medical costs	$375,394	$344,111	$316,121
Net nonmedical costs	$2,918	$2,228	$1,492
Total costs	$378,312	$346,339	$317,613
Transposition/DORV			
Net medical costs	$120,192	$113,606	$107,020
Net nonmedical costs	$4,562	$3,479	$2,323
Total costs	$124,754	$117,085	$109,343
Tetralogy of Fallot			
Net medical costs	$194,283	$179,465	$161,354
Net nonmedical costs	$5,800	$4,422	$2,954
Total costs	$200,083	$183,887	$164,308
Single ventricle			
Net medical costs	$227,212	$163,000	$123,485
Net nonmedical costs	$3,507	$2,674	$1,786
Total costs	$230,719	$165,674	$125,271
The costs presented in this chapter were current in the year the chapter was written. They can be updated using inflation factors accessible by clicking below.			
Link to inflation factors			

III.5.B.3 Other Studies

Several other studies have been conducted on the costs of cardiac abnormalities. In particular, two recent studies are especially useful for comparison. The first, by Pearson et al. (1991), looks at hospital use and inpatient charges for cardiac disease patients in the first year of life. The second, by Silberbach et al. (1993), predicts the hospital charges for congenital heart disease surgery. These two studies are easiest to compare to Waitzman et al. because they examine similar types of direct medical costs. Although these studies vary slightly in their methodologies, the results tend to corroborate the Waitzman et al. results. No recent studies examining the direct non-medical costs associated with cardiac abnormalities were identified.

III.5.B.3.1   Pearson et al. (1991)

The Pearson et al. study analyzes the inpatient charges for infants with cardiac disease in the first year of life. Infants admitted to The Johns Hopkins Hospital Children's Center in Maryland in 1988 were identified for the study. Complete data for 93 of these infants were available. The infants were subdivided into three groups: those with complex diseases, those with extra-cardiac anomalies, and those with both. Hospital charges were recorded, including all routine care charges, laboratory charges, medical and surgical supplies, physical therapy, operating room time and supplies, radiologic procedures, pharmacy supplies, and blood and blood related products.

Pearson et al. estimated hospital charges per infant. These charges have been adjusted to 1996 dollars using the Consumer Price Index for medical care (1996:1988=1.6465) to facilitate comparison across studies. Average hospital charges per infant were estimated at $60,506. The number is larger for infants with complex cardiac diseases.

Pearson et al.'s results are similar, in the first year of life, to those reported by Waitzman et al., particularly for transposition/DORV, tetralogy of Fallot and single ventricle (see Table III.5-1 for Waitzman et al.'s cost estimates). Waitzman et al.'s higher cost estimate for truncus arteriosus may be reflected in the upper range of costs that Pearson et al. estimated. They identified a range of $1,651 to $770,177 per patient.

III.5.B.3.2   Silberbach et al. (1993)

The Silberbach et al. study is also useful for comparison because it looks at the hospital charges for congenital heart disease surgery. Although the study is not strictly limited to children, and is on a per-surgery basis, 49 percent of the surgeries examined occurred in children less than 12 months old. The study is also useful because it breaks out costs according to ten different types of congenital heart disease, facilitating direct comparison with the Waitzman et al. study. The study was conducted between 1985 and 1989. A conservative adjustment to 1996 dollars assumes that all reported costs in the Silberbach et al. study were in 1989 dollars (Consumer Price Index for medical care (1996:1989=1.53)).

Silberbach et al. predicted the average hospital charge for a patient with congenital heart disease at $41,699. More specifically, they estimated that tetralogy of Fallot surgery costs $56,997 and surgery for transposition/DORV of the great arteries costs $44,789.

The Silberbach et al. study found similar costs to those reported in Waitzman et al., particularly in their finding that surgery for transposition/DORV is less expensive than for tetralogy of Fallot. The Silberbach et al. estimates are a bit lower. This difference in relation to Waitzman et al. and Pearson et al. is probably due to the limited nature of

the Silberbach et al. study; they calculate hospital charges for a specific surgical procedure and do not include other costs that would be incurred during the rest of the year.

III.5.C Conclusions

Because the Waitzman et al. study provides cost estimates past the first year of life, it is more appropriate for benefits evaluation than other studies reviewed. The other studies, discussed above, generally support the estimates provided by Waitzman et al.